

LAL and HSLipase Are Different Enzymes

- Different cell compartments
 - LAL in lysosomes
 - HSLipase in cytoplasm
- Different amino acid sequences
- Different genes
- Different post-translational modifications
- Different developmental/tissue expression
- Different functions in cell

Amino Acid Alignment (hLAL and h-HSL)

hLAL 83	10	20	30	
h-HSL 83				
	K M F L G L V C L W P - - - L H E S - - -			
	D L T M T Q S V T E D N I A F S G G E T A			
	M R			
hLAL 83	40	50	60	
h-HSL 83				
	Q R L S G V F A G V R E Q A L G L E P A L G R L L G V A . L			
hLAL 83	70	80	90	
h-HSL 83				
	T A N M S E I S Y W F P S E E Y E T E			
	F D P A Y R S V H T R C C L A H H K S			
	N . D P E T N . L .			
hLAL 83	110	120		
h-HSL 83				
	D C L N I P H G K N S D K G - - - - -			
	R - S N R S I F F T S N L A E L E A Y L A A L T			
	G Y . . R H			
hLAL 83	130	140	150	
h-HSL 83				
	- - - - - P - - - L Q - - H L			
	Q L R A L V Y Y A Q R L L V T . P G V . F G D G L			
hLAL 83	160	170	180	
h-HSL 83				
	S S N W V T N A N - - - - S S I L A D F D			
	F L R E Y V T H K R C Q F T P R P			
	A D L G C F Y G L G F A .			

190 210
hLAL 99 VWMGN R - - - - - NT S KHK T S Q D - -
h-HSL 99 FLQTI I - - - - - E H K K N E T G A A S - -
S GLV SFG R L S V S L

220 240
hLAL 99 - - - - -
h-HSL 99 FTSGRFAIDPELRGAEEFERITQNLDVHFWK

250 270
hLAL 99 E - - - - -
h-HSL 99 A FWNITEMEVLSSLANMASATVRVSRLLSL

280 300
hLAL 99 - - - - -
h-HSL 99 SYDEM KYD P NFI N - - -
EMPLT DPT T SPP A - - -
PPEAF A L I L TGP GP

310 330
hLAL 99 - - - - -
h-HSL 99 VLVRLLSYDLREGQDSE . . . S.GQR .

340 360
hLAL 99 T I I A F S E L A K R K M F P S V A
h-HSL 99 L E P R P Q R S R S L I F H F Q T S
Q P . . G V A

HLAL 03 370 F C T S 380 M K L R
h-HSL 03 R S H E P Y L K S W A Q E L G A P I S I D Y S A L P E P

HLAL 03 420 L D H I K D L 430 D K E F P Q S F K W L T
h-HSL 03 F R A E E C F Y C W A K H C L G S T E
P L F A L G . . C

HLAL 03 440 K E C 450
h-HSL 03 D S G T H . . G N L C F T V A L R A A Y G V R V P D G

HLAL 03 460 I M A A Y P A T M L Q P A A S P S R L L S L 470 L C G F N E R N
h-HSL 03 I M A A Y P A T M L Q P A A S P S R L L S L M D P L L P L S

HLAL 03 490 N M S R D T 500 P A G T S Q N L H W
h-HSL 03 L S K C S A Y G A K T E H S D N S D Q K L G M G L
V . Y G A K T E H S M

HLAL 03 520 S Q A V K F Q K F Q A D W S 530
h-HSL 03 V R R D T A L L R D F G S S W L N S F L E L S G R K

HLAL 03 550 570
h-HSL 03
- - - - - A K N Y F H Y N S Y P P
- - - - - S E A A L A Q P G P L G T
S Q K M S E P I A E P M R R S V Q

HLAL 03 580 600
h-HSL 03
Y N D M L P T V W S G H W L A V - - - - -
D S N L T R D S L R N S T S S T P E M S L S A

HLAL 03 610 630
h-HSL 03
- - - - - Y I T - - - - -
- - - - - S F P - - - - -
E T L S P S T P D V N L L P E D A G E E A E A K N E L

HLAL 03 640 660
h-HSL 03
- - Q I T N F E S I W E L D - - - - -
- - M D R G V A A F G F P R - - - - -
S P L . . P E H R S S Q G A T Q M P

HLAL 03 670 690
h-HSL 03
- - - - - I W G D W R L Y N - - - - -
- - - - - M S P L A P D S M L K S L P P V H I
L Y S S P I V K N P F

HLAL 03 700 720
h-HSL 03
- - - - - K I N M Y Q L
- - - - - D S M A L R
V A C A L D P M L D L R N L G Q P V T L R V

330

340

350

HLAL 33
h-h31 33

V E D L P H G F L T L A A L C R E T R Q A A E L C V E R I R

360

370

380